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Classic bladder exstrophy – the timing of initial closure and technical highlights

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The timing for the initial reconstruction of the bladder and abdominal wall in classic bladder exstrophy (CBE) has typically been performed in the immediate newborn period in both the modern staged repair of exstrophy (MSRE) and the complete primary repair of exstrophy (CPRE) protocols. The benefit of immediate surgical closure (typically in the first 72 hours) was that many children could be managed without resorting to osteotomy due to the malleability of the pelvis. The concerns about the potential short and longer-term impacts of osteotomy have waned mainly due to improvements in orthopaedic surgical management, the use of external fixation, and the ability to provide sedation and pain control to infants and children.[1,2] Additionally, an ongoing understanding of how to protect the bladder template and permitting a too-small-to-close bladder template time to grow also influences the reasoning behind the timing of initial bladder closure. Two other social factors have also led to delaying immediate reconstruction: the desire to provide time for maternal/parental-child bonding and the accessibility of an appropriate centre for care with experts in the management of exstrophy.

Keywords: classic bladder exstrophy, modern staged repair of exstrophy, pelvic osteotomy

Introduction

Delays in the initial closure until 3–4 months of age are now the norm for most infants in the United States. Though this represents a substantial change from historical teaching, there seems to be a consensus that initial bladder closure after the first 72 hours with the use of osteotomy and very secure one-month-long postoperative immobilisation provides equivalent successful outcomes. Unanticipated transfusion was the most common complication noted in an extensive series of closures performed after the first 30 days of life.[3] Studies on children undergoing closure at various time points in the first year of life seem to indicate a negative impact on bladder growth among those closed beyond nine months of age.[4]

The ideal timing for primary bladder closure is now considered to be in the first year of life, preferably before the ninth month. However, this timing is not possible in many parts of the world where patients may only present well beyond this ideal time, and available, intensive and immediate postoperative care for infants may be lacking. When closure is delayed, due to unforeseen or unchangeable circumstances, the primary goal should be to provide a secure bladder and abdominal wall closure at the first operation, as repeated attempts at closure further compromise long-term bladder growth.

Multiple techniques exist for the primary reconstruction of classic bladder exstrophy (CBE). The staged repair, initially described by Jeffs, has had the most extended clinical experience and continues to demonstrate a high degree of success.[5] Over the years, considerable changes have been made to the initially described approach and timing of the various parts of the staged reconstruction. In its current iteration, the modern staged repair of exstrophy (MSRE) proceeds in two (in females) or three (in males) separate surgeries: 1) initial secure abdominal wall and bladder closure (males and females); 2) epispadias repair before 24 months (males only); and 3) a definitive continence procedure using a Young-Dees-Leadbetter bladder neck reconstruction when the child can participate in an appropriate voiding programme (males and females). This modified staged approach has also been widely accepted and has had good outcomes in maintaining a secure abdominal wall closure, cosmetic and functional penile outcomes, and eventual continence.

Primary bladder closure and abdominal wall reconstruction

The initial step in the MSRE is the secure closure of the bladder and abdominal wall, which sets the stage for future procedures and, ultimately, urinary continence. In the MSRE, the birth defect is converted from a bladder exstrophy to a proximal epispadias. An intrinsic aspect of every stage of exstrophy management is a dedicated, well-trained, and highly knowledgeable team with expertise in all aspects of exstrophy management. This team includes paediatric urologists or surgeons, paediatric orthopaedic surgeons, paediatric anaesthesiologists adept in postoperative pain management, expert pre-, intra-, and postoperative nurses, psychologists or psychiatrists who have experience in managing complex medical conditions, and social workers who can assist the family through a very challenging multistage reconstructive process.

Preoperative evaluation and management

Preoperative preparation should include a comprehensive evaluation of the child's medical status, including determining normal blood counts and metabolic profiles. Ultrasound evaluation of the kidneys before closure helps demonstrate normal renal anatomy. Studies have shown that 2.8% of children with CBE have associated renal anomalies.[6] While not an exclusion criterion for staged reconstruction, knowledge of renal anomalies will aid postoperative fluid management and allow for ongoing renal outcome monitoring. Preserving renal function in CBE remains a key determinant of the success of reconstruction.[7] Where available, magnetic resonance imaging has been utilised to evaluate the pelvic floor and its changes that occur with successful reconstruction.[8, 9]

Bladder evaluation and protection

At birth or on initial evaluation, careful evaluation of the patient and the bladder template is crucial to determine the best time for surgical reconstruction. The bladder template is evaluated in the infant by carefully inverting the bladder template into the abdomen using a clean or sterile technique. Some templates may already present as inverted; in this case, there may be more bladder template evident on the exam than visually noted before the exam. If the exstrophied bladder template can be easily inverted well into the abdomen and pushed underneath the abdominal wall fascia with gentle pressure, the template is considered large enough to close. Another general rule of thumb is that the closed bladder should be able to fit around a 10 Fr Malecot catheter, which typically translates to at least a 3 cm diameter bladder template. The presence of polyps on the bladder template should also be noted, as these may be so numerous that they compromise the amount of template available for closure. Polyps were noted more commonly in patients with a delay in closure and were typically associated with squamous metaplastic change; however, no dysplasia was noted.[10] Resection of the polyps at the time of closure is appropriate.

A very small bladder template < 3 cm in diameter at birth is a reason to consider delaying closure. When closure was delayed in this circumstance, later closure with osteotomies and external fixation was very successful.[11] Despite the successful delayed closure, the time to continence was prolonged due to slower bladder growth, and most patients eventually required a catheterisation regimen using a continent channel (i.e. a Mitrofanoff) to be dry.[11] Other studies indicated that while a small bladder template will grow following closure, the eventual capacity achieved will still be smaller than those closures starting with a larger bladder template.[12] Initial surgical management closes the bladder and proximal urethra and places it deep in the pelvis, allowing for tension-free abdominal wall closure. This is best achieved with the use of osteotomy if the closure is beyond the first 72 hours of life.

Since immediate postnatal closure is becoming more infrequent, protection of the bladder template becomes more crucial to prevent damage and metaplastic changes to the bladder mucosa. This protection should start at birth by avoiding large plastic clips to clamp the umbilical cord. A more suitable alternative is to use silk ties to ligate the umbilical cord; this will prevent damage to the bladder template. Additionally, the bladder template must be covered with a protective barrier. Using saline to moisten the bladder template and non-adherent thin plastic wrap will prevent drying of the mucosa and keratinisation. Alternatives to plastic wrap include a gauze saturated in petroleum-based ointment. An uncovered bladder template can be very sensitive and painful and is subject to metaplastic changes.

Good parental preparation is critical to ensure families understand the extent of the surgical undertaking and the lengthy postoperative course typically required for success. Additionally, having good family and social support plays an integral part in the eventual surgical and clinical success of the child with exstrophy.

Intraoperative management

An experienced operative team is essential for the eventual successful outcome. Strict attention to detail remains a hallmark of success and includes the accurate placement of postoperative drainage catheters and the exact placement of epidural catheters and osteotomy pins. Access to intraoperative fluoroscopic imaging is helpful for the successful placement of epidural catheters and rapid evaluation of pin location during osteotomy. All patients are managed with a combined general and epidural analgesia approach.[13] Placement of central intravenous access is beneficial in the immediate postoperative phase to provide fluids, antibiotics, and pain medications, particularly when the patient is not receiving oral intake. The placement of central lines is also made easier by using intraoperative fluoroscopy.

The entire lower aspect of the child below the chest is prepped into the operative field. This permits all aspects of the procedure to be performed without repreparation and draping. Osteotomies are performed, and pins are placed in the various fragments following the osteotomy. The bladder template and posterior urethrovesical junction can then be dissected and closed. The bladder template and paraexstrophy skin flaps are identified by their mucosal-like or pinkish appearance relative to the normal skin tone lateral to the bladder plate and urethra. The umbilical stump is typically included in the initial dissection. This stump can anchor a holding stitch or instrument to aid exposure and provide traction; the stump is transected before bladder closure.

Below the skin, the surgeon should take care to identify the rectus fascia (which will be diastatic), the peritoneum, and the right and left umbilical arteries. The most technically challenging component is the separation of the intersymphyseal bands; their complete dissection is crucial to ensure that the posterior urethra/vesicourethral unit can be placed deep in the pelvis. The intersymphyseal bands are identified as shiny white tissue originating from the pubic symphysis and passing medially toward and inserted on the epispadiac posterior urethra and bladder neck. Failure to release these bands could lead to bladder prolapse or complete wound dehiscence, because the vesicourethral unit is still tethered anteriorly by these bands. The bands' colour and texture appear almost like fascia. A surgical tip to evaluate whether the intersymphyseal bands are appropriately dissected is for the surgeon to run their finger along the medial aspect of the pubis. More dissection is required if there is a tight, distinct piece of fibrous tissue.

However, extreme care and a slow, deliberate and measured dissection are absolutely required as the surgeon approaches the inferior aspect of the pubic symphysis. This is because the corpora cavernosa are attached to the inferior aspect of the pubic ramus; failure to identify the corpora could result in ischemic injury to the penis. Given the bony pelvic abnormality in exstrophy, the corpora

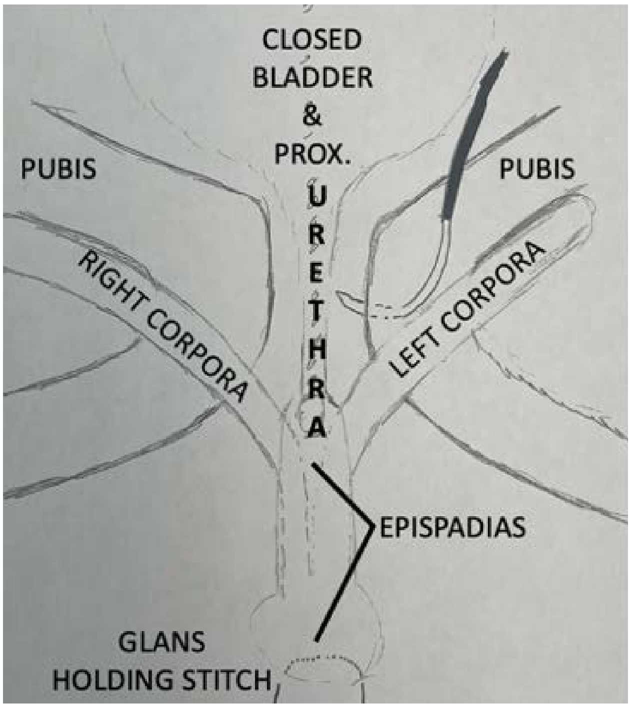

Figure 1: Anatomy before pubic closure

are more anterior or closer to the skin surface than typical. Their rounded, rod-like structure and soft feel are distinct from the shiny, white intersymphyseal bands. It should also be noted that at the time of bladder closure using the MSRE, the bladder neck does not undergo any reconstruction or tapering as is frequently done in the complete primary repair of exstrophy (CPRE).

Approximation of the pubic bones is best made with a No. 2 nylon-modified horizontal mattress suture on a large tapered needle.[14] Erosion of the intrapubic stitch into the soft tissues is possible, but this can be minimised by using a modified mattress-type suture placement (Figures 1 and 2). A large, tapered needle should be used, and the suture passed to keep it and the knot on the anterior surface of the pubis.

An off-the-shelf regenerative tissue matrix (such as AlloDerm RTM, Allergan Aesthetics, an AbbVie Company, Lake Bluff, Illinois, United States) can be used, if desired, as an additional protective layer between the closed urethra and bladder. This serves to reduce the possibility of stitch erosion into the soft tissues. A small rectangle of the regenerative tissue matrix is laid over the closed proximal urethra and bladder neck and sewn to more lateral soft tissue. Three surgeons are needed to tie the pubic stitch successfully. One surgeon places their index fingers on the greater trochanters and their thumb on the pubis/pubic rami and rotates the pelvis anteriorly and medially, thereby reducing the pubic diastasis. A second surgeon exposes the apposed pubis and removes any fluid with suction. The third surgeon ties the knot. Tying the pubic stitch introduces a slight bend in the corpora cavernosa, which places them at risk for venous congestion. The surgeon must observe the penis. If it stays ischaemic, the pubic stitch should be removed, and a fresh stitch placed more anteriorly in the pubis, which moves the stitch farther away from the corpora, reduces their bend, and lowers the risk of corporal/penile loss.

Ureteral stents and suprapubic tubes maintain bladder drainage, which is important for the stability of the bladder and abdominal wall closures. Ensuring that the tubes and stents are in excellent position and draining well before the procedure is completed lessens the concern for postoperative problems caused by inadequate drainage.

Combining bladder closure and epispadias repair is considered in select older boys. While this is done as a standard component of the "complete" repair (also referred to as the CPRE) as described by Grady et al.,[15,16] it has also been successfully used in the context of the staged reconstruction in select older boys. Epispadias repair is performed using the Cantwell-Ransley technique.[17,18] Limiting the use of this technique (i.e. combined bladder closure and epispadias repair) to older boys has avoided the issues of corporal and possibly penile loss, a devastating and irreversible complication seen with other techniques.[19,20]

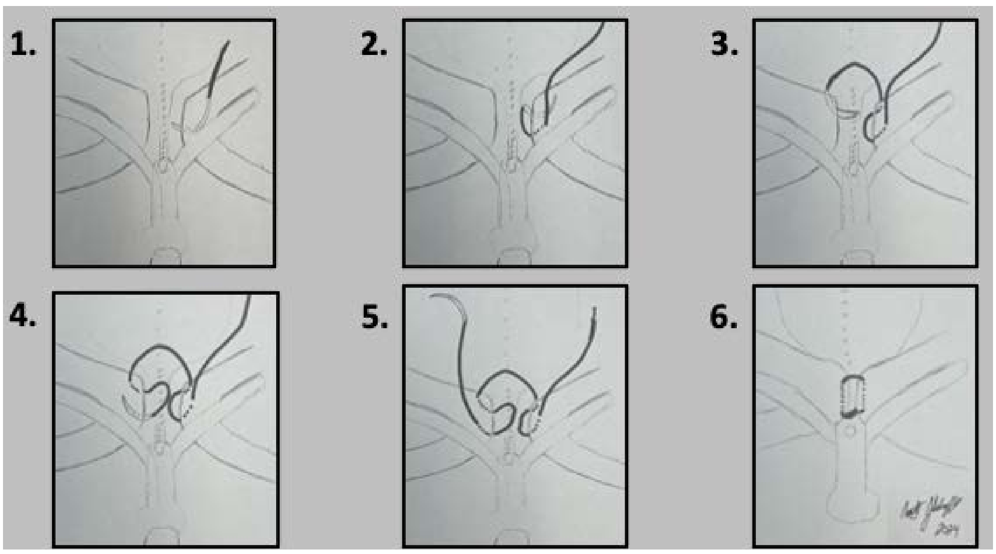

Figure 2: Placement of the pubic stitch; a modified horizontal mattress stitch using a No. 2 nylon on a large tapered needle is placed to lower the risk of stitch erosion into the urethra or bladder, no stitch crosses directly over the closed bladder/urethra and the knot is on the anterior side of the closed pubis

Table I: Important considerations regarding primary bladder exstrophy closure

1. Historical teaching suggested that all bladder exstrophy closures should be urgently performed postnatally within the first 72 hours. When closed within this timeframe, pelvic osteotomies are often not required.
2. Today, delayed primary bladder exstrophy closure (within the first nine months of life) is the new norm in the United States.
3. Delayed closure advantages include parent-child bonding and easier scheduling of the procedure. However, the main disadvantage of delayed primary bladder closures is that they uniformly require pelvic osteotomies, which adds additional morbidity.
4. Key manoeuvres that promote the success of the initial bladder closure include: • Complete dissection of the intersymphyseal bands. The inferior portion of the bladder template (located near the bladder neck) must be completely dissected from these shiny, white, fibrous bands to allow the vesicourethral unit to move deep into the pelvis. Failure to fully release the bands is one reason for bladder prolapse and failed initial repair. • Optimal and complete urinary drainage with ureteral catheters and suprapubic tubes. • Pelvic osteotomy when closure occurs > 72 hours after birth. Without pelvic osteotomy, there is substantial tension on the soft tissue closure, another reason for failed initial repair. • Adequate postoperative immobilisation and a sufficient pain control regimen that keeps the patient still and comfortable.
5. The following healthcare providers are crucial components of a multidisciplinary exstrophy team: • Paediatric anaesthesia (adept in postoperative pain management) • Paediatric orthopaedic surgeon • Paediatric urologist or surgeon • Postoperative nurses experienced in recognising and mitigating postoperative problems • Paediatric psychologists or psychiatrists • Social workers

Postoperative management

Postoperative traction and immobilisation with an external fixation device are instituted at the end of the operative procedure. Care is required during the patient transfer out of the operative theatre to ensure that the pelvis remains stable for 4–6 weeks. Traction is performed using a modified Buck's technique, as described by Wild et al.[21] Intermittent radiologic evaluation is performed beginning at 4 weeks to confirm that pelvic healing is progressing. Transfusion may be required to accommodate for blood loss from the osteotomies.[22,23] The reader is directed to other manuscripts in this journal edition for more details on pelvic osteotomies and multimodal postoperative pain management.

Successful postoperative management also includes reducing abdominal distension using nasogastric drainage to decrease tension on the abdominal wall closure. Central venous access will permit hydration in the immediate postoperative period and can be used for parenteral nutrition in patients with a delayed return of bowel function. It can also be critical in providing pain control in the event of early dislodgement of epidural catheters.

The success of postoperative management depends on well-trained nursing staff with experience in administering care and recognising and troubleshooting problems in exstrophy patients after their initial closure. The first 72 hours are best managed in an intensive care setting (i.e. with a 1 : 1 nurse-to-patient ratio) with a gradual reduction in intensive care needs after the patient's pain control regimen becomes apparent, bowel function returns, and their overall medical situation stabilises. The nurses' ability to rapidly identify and successfully act upon problems, such as a dusky appearance of the penis, the lack of drainage from ureteral and suprapubic catheters, significant patient discomfort leading to increased movement of the pelvis, or noticeable increases in abdominal girth (from possible ileus), are critical to the ultimate success of the operative procedure and cannot be minimised.

Failure of reconstruction

Multiple factors have been identified that lead to the failure of reconstruction. These include intraoperative factors such as inadequate dissection of the intersymphaseal bands and pelvic floor leading to superficial placement of the bladder and posterior urethra and a lack of good bladder drainage with appropriate stents and tubes.[24,25]. Arguably, the most critical factors in the failure of initial reconstruction seem to be the lack of good pelvic fixation and the use of osteotomies.[26]

Failure of reconstruction has significant short- and long-term impacts on the patient and family. Failure of initial reconstruction has been associated with a decreased potential for bladder growth and voided continence.[27,28] Although reclosure with osteotomies has been shown to have reasonable success rates, successful initial closure remains a crucial first step for attaining functional success in CBE reconstruction (Table I)

Conclusion

Multiple factors determine the timing of initial surgical reconstruction for bladder and abdominal wall closure. Immediate newborn closure, while reducing the need for osteotomy, may not be easily achievable. Delaying closure may have the benefit of allowing parental bonding and providing care at a centre of excellence. Improvements in the surgical management of CBE, have provided excellent surgical and functional outcomes that can be reproduced in most clinical settings.

Conflict of interest

The authors declare no conflict of interest.

Ethical approval

The authors declare that this submission follows the principles laid down by the Responsible Research Publication Position Statements developed at the Second World Conference on Research Integrity in Singapore in 2010. This narrative review/study is exempt from Institutional Review Board approval because it did not involve human subjects.

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